Prenatal diagnosis of Galen Vein aneurysm: when to deliver?

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Galen vein aneurysm is a rare occurrence involving cerebral vessels and leading to a high output cardiac failure, with ultimately fetal demise. We report a case that highlights the difficulty in clinical management of this entity.

A 23 year old G1P0 spontaneous pregnancy was referred at 28 WA for a Galen Vein aneurysm. Ultrasound finding include a 35 by 14 mm vascular structure with a turbulent flow occupying the central part of the head. Biometry is around the 20th percentile. Cardiac overload was evident with a spherical looking heart, dilated neck vessels (white arrow) and tricuspid regurgitation. At 33 weeks, IUGR was more obvious, reaching the 10th percentile in all sites. Umbilical artery doppler showed then an absent diastolic flow, ductus venosus had an abnormal a wave. Midcerebral artery Doppler was measurable despite the aneurysm and was still in the normal range. After 48 hours in-hospital surveillance by non-stress test twice daily she had a cesarean section because of a persistent non reactive fetal heart rate, with absent variability. Baby weighed 1800 g and was doing relatively well with an Apgar score of 9 at 5 minutes. As the baby was stable for the first 48 hours, occlusion of the aneurysm by interventional radiology was postponed until a weight increase to about 2500 g. This decision was based on literature review, expert opinions in centers having a relatively large experience in cases of Galen vein aneurysm, and also based on the preference of our radiology team. Unfortunately despite a stable state the first ten days, cardiac function deteriorated abruptly on day 11 and the baby died of heart failure despite medical management. This case raises the question about the timing of delivery and the timing of interventional radiology, with both prenatal and postnatal outcomes being at risk for these babies.

Prognosis depends on gestational age at diagnosis and aneurysm size. Some cases show regression following delivery, suggesting a more conservative approach after delivery. In our case the relatively favorable postnatal outcome led after 11 days to a rapid deterioration of the cardiac status. A similar case is reported by Cherif et al who found that after an initial period of improvement cardiac status deteriorated suddenly at day 14, which led to fetal death at day 36 despite medical treatment; authors conclude that heart failure can occur long after the first days of life. Embolization is the therapy of choice; the results are variable as some authors report high failure rate and neonatal death. Outcome seems poor, with only about 30% of patients diagnosed with the condition alive without mental impairment. Deciding for delivery to allow radiology intervention seems to have a limited impact because of the already serious prognosis involved and the timing of this decision can be challenging.